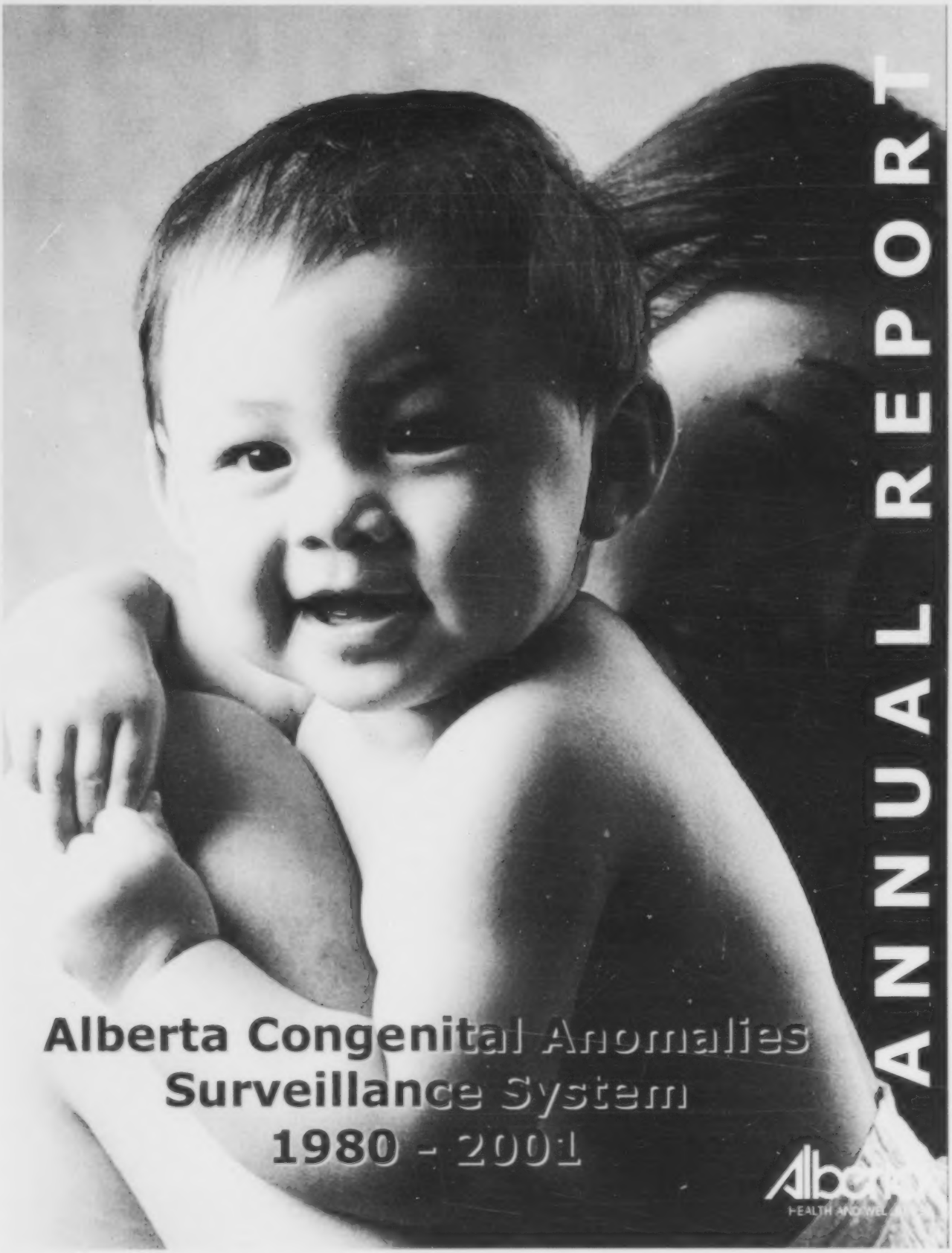
ANNUAL REPORT
Alberta Congenital Anomalies
Surveillance System
1980 - 2001

105-04234/

ALBERTA CONGENITAL ANOMALIES SURVEILLANCE SYSTEM

SIXTH REPORT

1980 – 2001

Alberta Children's Hospital Research Centre
Department of Medical Genetics
University of Calgary
1820 Richmond Road SW
Calgary, Alberta T2T 5C7

Division of Vital Statistics
Alberta Registries
10365-97 Street
Edmonton, Alberta T5J 3W7

Alberta Health and Wellness
Health Surveillance
PO Box 1360 STN MAIN
Edmonton, Alberta T5J 2N3

Medical Consultant: R.B. Lowry
Manager: B. Sibbald
Health Surveillance Consultants: L. Svenson
F-L. Wang

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For additional information please contact:

Health Surveillance
Alberta Health and Wellness
P. O. Box 1360 STN MAIN
Edmonton, AB T5J 2N3
CANADA

Telephone: +1-780-427-4518
Toll-Free: 310-0000 (Alberta only)

Website: www.health.gov.ab.ca

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The success of ACASS depends upon the interest and activities of many people. We thank health records personnel in the province's hospitals, unit clerks, nurses, clinic co-coordinators and physicians. Many physicians were contacted by letter in order to obtain additional clarifying information and their prompt replies are appreciated. The additional information is vital in improving the quality of the data. We thank the Hospital Systems branch and Vital Statistics staff for expediting documents.

We are housed in space associated with the Department of Medical Genetics at the Alberta Children's Hospital, Calgary Health Region. We would like to thank the Department and the Calgary Health Region for their continuing support.

Finally, a special thanks to our clerical staff Anne Preece and Judy Anderson, our research assistant Tanya Bedard and our occasional medical consultants.

Medical Consultants

Principal: R.B. Lowry, MD, DSc, FRCPC, FCCMG
Occasional: J. Harder, MD, FRCPC, Paediatric cardiologist
R. Sauvé, MD, FRCPC, Community Health Sciences
C. Trevenen, MD, FRCPC, Paediatric Pathologist

Health Surveillance, Alberta Health and Wellness

S. Gabos, MD, Director
F-L Wang, MB, PhD, Epidemiologist
L. Svenson, Team Lead, Epidemiologic Surveillance

ACASS Staff

B. Sibbald, MSc, Manager
T. Bedard, BSc, Research Assistant
J. Anderson, Secretary
A. Preece, Clerical Assistant

Alberta Registries, Vital Statistics

G. Brese, System Administrator
B. Haugrud, Assistant Director

EXECUTIVE SUMMARY

1. This is the sixth in a series of reports detailing the prevalence of congenital anomalies in the province of Alberta. Aggregate data is also included from 1980 onwards. The main emphasis is on Section XIV (Congenital Anomalies) of the *International Classification of Diseases, 9th Revision* (ICD-9). We do monitor selected items from other sections however, such as disorders of metabolism. Data on such disorders will be provided to interested parties upon request.

2. This is the last report to use the ICD-9 coding system. The new *International Classification of Diseases, 10th Revision* (ICD-10) classification system has now been adopted in Alberta for congenital anomaly reporting so future reports will use ICD-10.

3. As noted in the previous report, the overall frequency of most congenital anomalies remains relatively unchanged with the exception of neural tube defects that continue to decline. The decline is most obvious with anencephaly, although there was a significant downward trend with spina bifida as well. Although some of the decline might be attributable to termination of pregnancy, folic acid fortification may also be playing a role.

4. The percentage of births to women 35 years of age and over has stabilized over the past three years. About 14.5 per cent of women 35 years of age and over gave birth in the period 1999-2001.

5. Limb reductions do seem to be increasing overall and we will be undertaking a review of these anomalies in the coming months. As noted in the last report, this is a very heterogeneous category and can range from the absence of part of a finger to a missing arm or leg.

6. Abdominal wall defects overall are not increasing significantly. However, when one examines the defects included in the category, an increase in the rate of gastroschisis is noted. On the other hand, omphalocele rates have been very stable over the years. The occurrence of gastroschisis is more frequent in younger mothers in Alberta, which is consistent with observations from other jurisdictions.

7. Because of some concern over the frequency of anophthalmia/microphthalmia, a detailed review of ACASS data was undertaken. The review showed that as an isolated defect, it is very rare and not increasing. The majority of cases were associated with a syndrome especially Trisomy 13 (Patau Syndrome).

8. Since there are substantial differences between ACASS and the Canadian Congenital Anomalies Surveillance System (CCASS) for anorectal atresia/stenosis, a detailed review has been started. Preliminary results indicate that a substantial number of cases belong in the multiple congenital anomaly categories such as VATER/VACTERL.

9. ACASS is a member of the recently established Canadian Congenital Anomalies Surveillance Network (CCASN), a Health Canada initiative, and is part of the Advisory Group for the network. The network has been formed to support the development, and maintenance, of high quality population based surveillance systems of congenital anomalies.

10. ACASS is participating in a national study on the impact of folic acid on the rates of neural tube defects for the years 1993-2002. The study is funded by the Canadian Institutes of Health Research (CIHR) through 2006 and will include data from Newfoundland, Nova Scotia, Prince Edward Island, Quebec, Manitoba, Alberta and British Columbia.

11. ACASS has been an associate member of the International Clearinghouse of Birth Defects Monitoring System since 1996 and participates in on-going collaborative studies.

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INTRODUCTION

History

The history of the Alberta Congenital Anomalies Surveillance System (ACASS) has been well described in previous reports. Although we have had some difficult years, our funding has been stable since 1996, when Alberta Health and Wellness, Health Surveillance Branch, assumed financial responsibility. We also work closely with Alberta Vital Statistics, as we have done since 1980, and rely on them to provide us with notifications of births, deaths and stillbirths.

Purpose of a Surveillance System

Public health surveillance is the ongoing, systematic collection, analysis and interpretation of data (e.g. regarding agent/hazard, risk factor, exposure, health event). These data are essential to the planning, implementation and evaluation of public health practice, closely integrated with the timely dissemination of these data to those responsible for prevention and control.

The purpose of surveillance for congenital anomalies (CAs) is to:

- provide reliable and valid baseline data of congenital anomalies in Alberta;
- investigate any significant temporal or geographic changes in the frequency of congenital anomalies with a view to identifying environmental, and therefore, possible preventable causes;
- measure trends;
- assess the effectiveness of prevention (e.g. folic acid or antenatal screening);
- assist with health related programme planning and development through the provision of data.

As well, it is important to look at patterns or associations of malformations to determine whether they belong to an existing or new syndrome complex. A principal feature of a surveillance system is timeliness. However, data collection and analysis should not be accomplished at the expense of accurate diagnosis.

Definitions

A **congenital anomaly** is an abnormality that is present at birth, even if not diagnosed until months or years later. Most congenital anomalies are present long before the time of birth, in the embryonic period (up to the end of the 7th week of gestation) and others in the foetal period (8th week to term). The term "anomaly" covers all the major classes of abnormalities of development, of which there are four major categories as follows:

Malformation – a morphologic defect of an organ, part of an organ or a larger region of the body resulting from an intrinsically abnormal developmental process (e.g. spina bifida, cleft lip and palate).

Deformation – an abnormal form, shape or position of a part of the body caused by mechanical forces (e.g. extrinsic force such as intrauterine constraint causing some forms of clubfoot).

Disruption – a morphologic defect of an organ, part of an organ or a larger region of the body resulting from the extrinsic breakdown of, or an interference with, an originally normal developmental process (e.g. an infection such as rubella or a teratogen such as thalidomide).

Dysplasia – the abnormal organization of cells into tissues and its morphologic result (e.g. Marfan Syndrome, osteogenesis imperfecta).

Ascertainment

An infant can be ascertained at any time up to the first birthday. Multiple ascertainment of the same infant can occur and indeed, are encouraged, as this frequently improves the quality and reliability of the data.

As several malformations may occur in the same infant, it is desirable to allow each to be reported, so that groups of associated malformations may be studied. This, however, leads to difficulties, as the final tabulations may be reported as total malformations (anomaly rates) or as the total number of malformed infants (case rates).

ACASS obtains information about infants with congenital anomalies from a variety of independent sources. Acquisition of additional reporting agencies is always a priority, as the use of multiple sources of information improves both the ease and completeness of ascertainment as well as the accuracy of the diagnostic data.

ACASS screens many important Alberta Health and Wellness and Alberta Vital Statistics documents for the presence of a congenital anomaly. These documents include:

- *Notice of a Live birth or a Stillbirth* often referred to as the Physician's Notice of Birth (PNOB)
- Medical Certificate of Stillbirth
- Medical Certificate of Death

All acute care hospitals in the province notify Alberta Vital Statistics of the live birth, stillbirth, admission or hospital death of an infant less than one year of age who has a congenital anomaly. A notification form called the Congenital Anomaly(ies) Reporting Form (CARF) is completed by the hospital health records personnel following the birth or an admission of an affected child. This form serves as the single most important source of case ascertainment.

Since many children with congenital anomalies are not admitted to hospital, it is important to obtain out-patient information from the Calgary and Edmonton Departments of Medical Genetics.

Ascertainment at a continued high level requires each hospital record department and each health care provider to co-operate with the system by notifying us as promptly as possible. We are fortunate in having such co-operative agencies and personnel.

Quality Control Measures

When a copy of a reporting document reaches the ACASS office in Calgary, it is scanned for content by the research assistant and manager. If the information is unclear, such as a vague or queried diagnosis, the manager, on behalf of the medical consultant, writes to the physician responsible for the case seeking clarification. A stamped, addressed envelope is included with the letter and the physician is asked to respond at the bottom of the letter, thus making the mechanics of replying easy.

The response from physicians has been very satisfactory (greater than 90 per cent) and usually this is sufficient to make a decision whether to accept or reject an anomaly or case. Any questionable diagnosis that is not confirmed is not entered into the database. Some cases are not included, such as those containing diagnoses that do not belong in a congenital anomaly system or are part of a normal developmental process. This could include patent ductus arteriosus or undescended testes in a premature infant. Any reports requiring a medical decision are reviewed with the medical consultant. Policy decisions with respect to the acceptance or rejection of a case and its coding are referred to the ACASS Advisory Committee. This body is comprised of a paediatric cardiologist, neonatologist/epidemiologist, paediatric pathologist and medical geneticist (medical consultant) with occasional input from a paediatric neurologist, paediatric nephrologist and a paediatric orthopaedic surgeon.

Coding

Coding is done at the Calgary office using the British Paediatric Association (BPA) adaptation of the *International Classification of Diseases, ninth edition* (ICD-9). We are currently also using the Royal College of Paediatrics and Child Health (RCPCH) adaptation of the recently introduced *International Classification of Diseases, tenth edition* (ICD-10). Difficult cases are referred to the medical consultant (medical geneticist). In the past, we were able to code only six anomalies, but since 1997 we have been coding all eligible anomalies reported to us.

Confidentiality and Release of Data

Notifications of congenital anomalies are sent to Health Surveillance, Alberta Health and Wellness, and then to the ACASS office in Calgary where the database is maintained. The notifications are handled by the manager, research assistant, secretary and medical consultant only. The data are treated in a completely confidential manner and the notifications are kept in locked files in a locked room. The database is secured by limited access and is password protected. Names are not disclosed to anyone. Should further clarification about a case or anomaly become necessary, we communicate with the attending physician or physician responsible for ongoing care. Direct contact is never made with the family. When data are requested, they are released in aggregate form with no personal identifiers.

Methodology and Limitations

Unless otherwise stated, the birth defect rates presented in this report are calculated using the following formulae:

$$\text{Anomaly (defect) Rate} = \frac{\text{Number of a given congenital anomaly among live births and still births}}{\text{Total number of live births and stillbirths}} \times 1{,}000$$

$$\text{Case Rate} = \frac{\text{Number of individual infants (liveborn or stillborn) with} \geq 1 \text{ congenital anomaly}}{\text{Total number of live births and stillbirths}} \times 1{,}000$$

Confidence intervals (approximate 95 per cent) are also included because the rate obtained is actually only a point estimate of the unknown, true population rate. The confidence interval provides information about the precision of the estimate. Thus, the confidence intervals are an estimated range of values where there is a 95 per cent probability that the true population rate will fall.

One of the major limitations of the surveillance system is that on its own, the information provided to us does not allow studies to determine aetiology. If increasing trends indicate there is a potentially serious problem, then separate investigative studies will need to be done. However, it is possible to conduct linkage studies with other data sources to explore potential causes of specific birth defects.

DATA

Trends

The following table and graphs of selected sentinel anomalies indicate the trends in congenital anomaly rates in Alberta from 1980 through 2001. Sentinel anomalies are those which the International Clearinghouse of Birth Defects Monitoring Systems watches worldwide, the rationale being they are quite easily identified and therefore will be accurately reported.

Table 1. Chi Squared Linear Trend Analysis and p-values for Selected Anomalies 1980-2001 Inclusive (Live Births and Stillbirths)

Anomaly	Trend Direction	Chi Squared Analysis (χ^2_{LT})	p-value
Neural Tube Defects	Decreasing	19.89	0.0000
Anencephaly	Decreasing	24.26	0.0000
Spina Bifida	Decreasing	5.03	0.0249
Hydrocephalus	No significant change	3.31	0.0689 (n.s.)
Cleft Lip +/- Cleft Palate	No significant change	0.60	0.4386 (n.s.)
Cleft Palate	Increasing	7.89	0.0050
Oesophageal Atresia/Stenosis	No significant change	1.65	0.1990 (n.s.)
Anorectal Atresia/Stenosis	No significant change	2.54	0.1110 (n.s.)
Hypospadias and Epispadias	No significant change	0.30	0.5839 (n.s.)
Limb Reductions	Increasing	12.44	0.0004
Abdominal Wall Defects	No significant change	3.45	0.0633 (n.s.)
Gastroschisis	Increasing	5.23	0.0222
Omphalocele	No significant change	0.04	0.8412 (n.s.)
Down syndrome	Increasing	17.22	0.0000
Renal Agenesis	Increasing	4.54	0.0331
Hypoplastic Left Heart Syndrome	No significant change	0.89	0.3455 (n.s.)

*Hypospadias and Epispadias calculated for male live births only

Neural Tube Defects

Neural tube defect (NTD) rates continue to decline in Alberta. Spina bifida rates are starting to fall somewhat, but the overall decline in NTDs can be attributed more strongly to the marked decrease in anencephaly rates. Terminations of affected pregnancies might account for part of the decline, but it is unlikely that this factor alone explains the total reduction of NTDs in Alberta. We have been able to follow terminations of pregnancy (ToPs) from 1997 only. However, if we extrapolate our ToP data through the years, there seems to be a true decline in the rates of NTDs since the 1998 introduction of folic acid fortification in flour and cereal/grain products (150 µg/100 gm). We have submitted a paper for publication outlining our observations. We are also participating in a Canada wide study of the impact of folic acid fortification and the prevalence of neural tube defects.

Figure 1. Rate per 1,000 births of neural tube defects in Alberta, 1980 to 2001

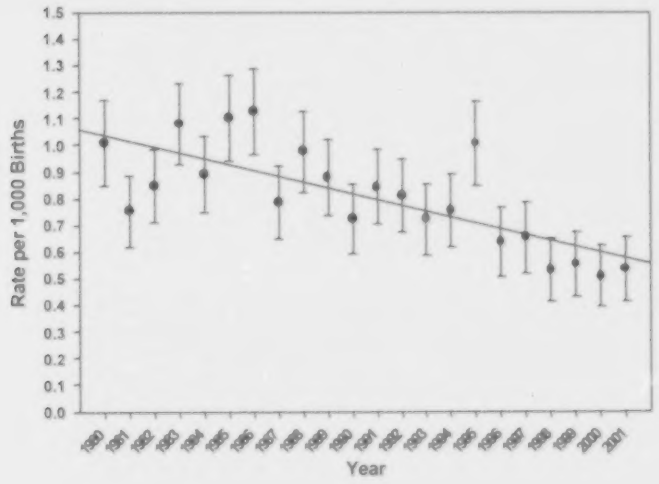

Figure 2. Rate per 1,000 births of anencephaly in Alberta, 1980 to 2001

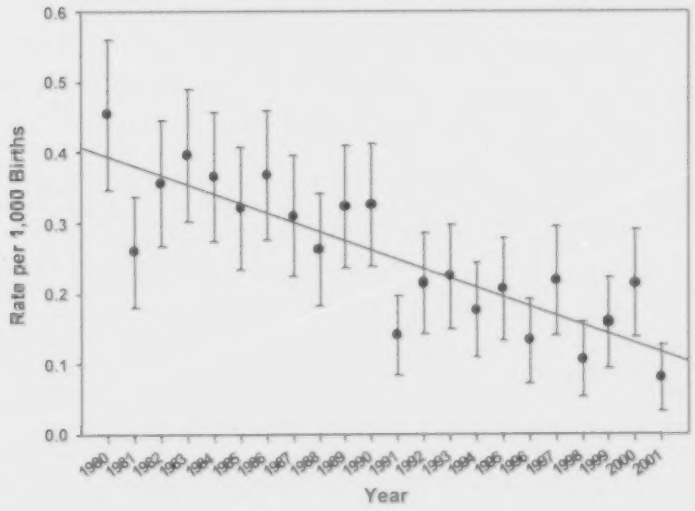

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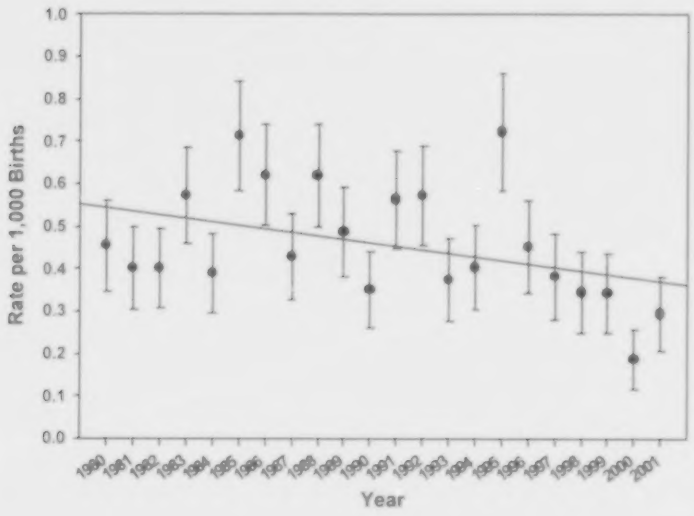
Figure 3. **Rate per 1,000 births of spina bifida in Alberta, 1980 to 2001**



Cleft Lip and Palate

The birth prevalence of cleft lip with or without cleft palate (CL ± CP) remains stable. There does appear however, to be a slight upward trend in the rates of cleft palate alone (CP).

Figure 4. **Rate per 1,000 births of cleft palate in Alberta, 1980 to 2001**

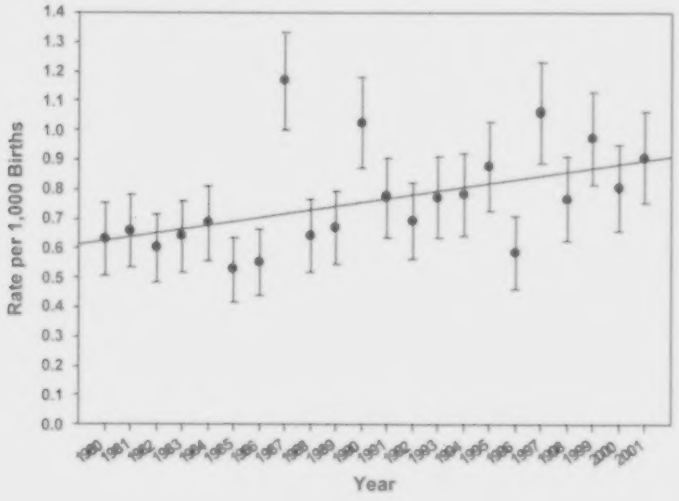
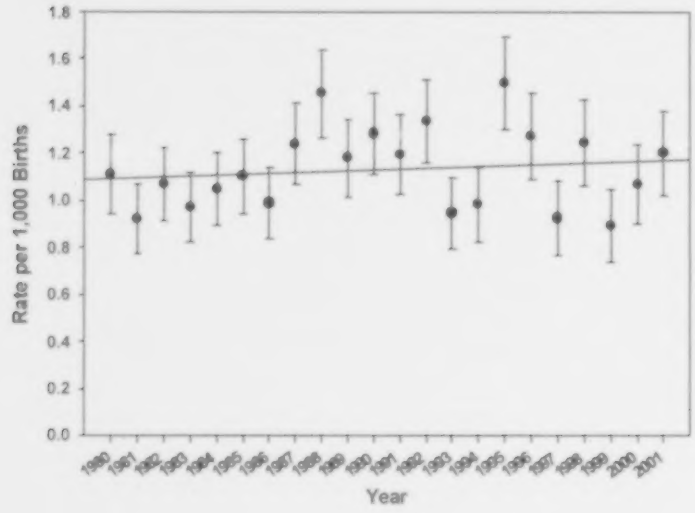


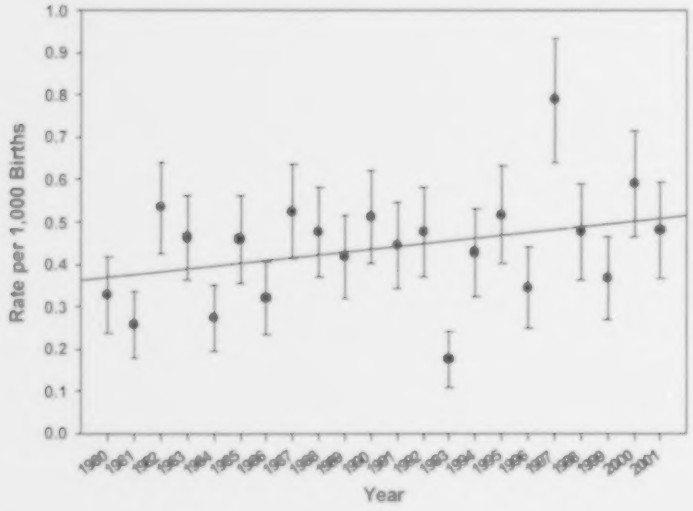
Figure 5. Rate per 1,000 births of cleft lip +/- cleft palate in Alberta, 1980 to 2001



Abdominal Wall Defects

Abdominal wall defects include mainly gastroschisis and omphalocele. Although there is no statistically significant increase in the rates of abdominal wall defect overall, the *apparent* increase can be attributed to the rising rates of gastroschisis. Omphalocele has remained stable over the years.

Figure 6. Rate per 1,000 births of abdominal wall defects in Alberta, 1980 to 2001



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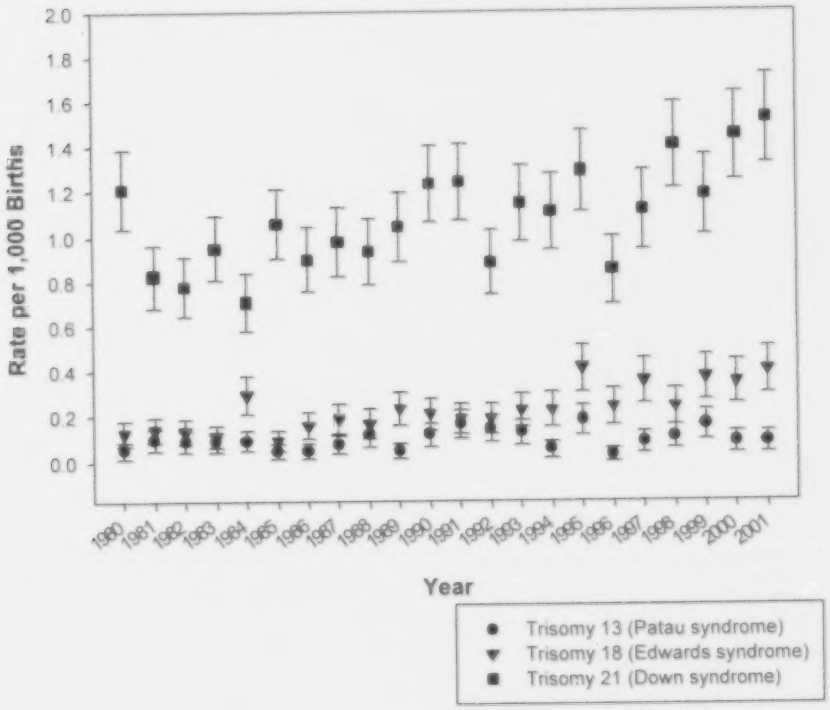
Chromosome Anomalies

From 1980-2001 there were 1,791 chromosome anomalies reported to ACASS. Of these, 1,366 or 76 per cent were either Trisomy 13 (Patau syndrome), Trisomy 18 (Edwards syndrome) or Trisomy 21 (Down syndrome). Down syndrome was the most commonly ascertained chromosome anomaly – 77 per cent of the above mentioned group of trisomies and 58 per cent of the total number of chromosome anomalies reported. Sex chromosome anomalies accounted for approximately 11% of the total.

As previously reported, Down syndrome rates are increasing (Figure 7). In most instances, Down syndrome is associated with increasing maternal age.

Figure 7. Rate per 1,000 births of chromosome anomalies (Trisomy 13, Trisomy 18, Trisomy 21) in Alberta, 1980 to 2001



Maternal age at delivery seems to be increasing overall in Alberta (Figure 8), in particular the percentage of babies born to women over 35 years of age
(Figure 9).

Figure 8. Maternal age distribution of live births

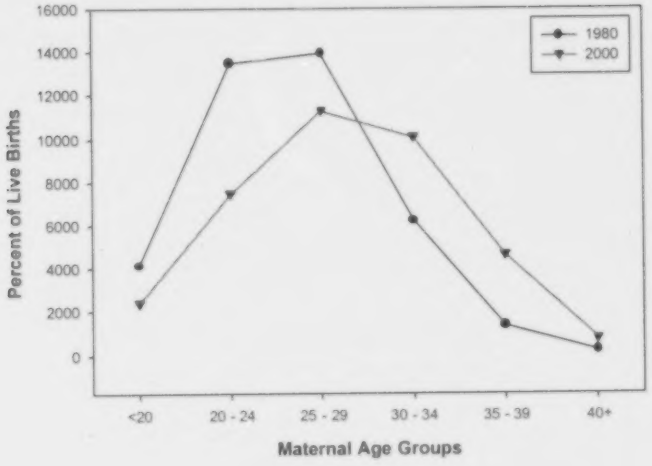
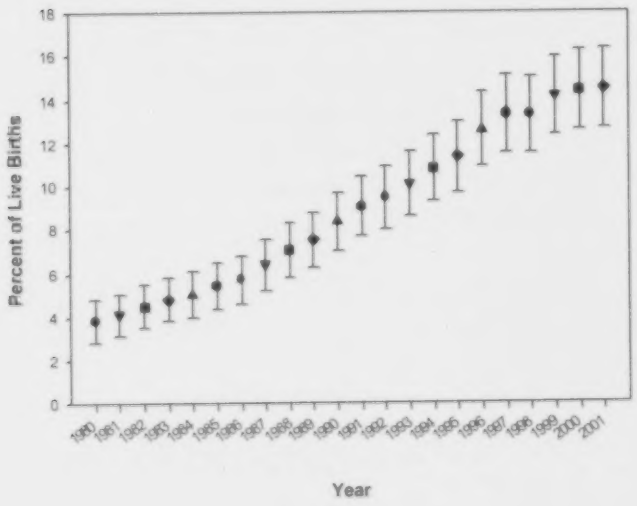


Figure 9. Percent of live births to women ≥ 35 years of age



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Terminations of pregnancy (ToPs) do not affect rates of Down syndrome greatly until we examine births to women over the age of 30. ACASS has collected data on ToPs since 1997. Table 2 illustrates the rates of Down syndrome by maternal age for the years 1997 to 2001. The rates including ToPs are in brackets.

Table 2. Down syndrome rates per 1000 total births by maternal age, 1997-2001

Maternal Age	Year				
	1997	1998	1999	2000	2001
<20	0.00	1.51	1.53	0.41	0.00 (0.43)
20-24	0.81	0.52	0.25	0.53	0.26 (0.40)
25-29	0.51	1.27 (1.36)	0.60	1.32	1.39
30-34	1.78 (1.88)	0.76 (0.95)	1.26 (1.55)	1.78 (1.98)	1.52 (1.70)
35-39	2.11 (3.51)	3.89 (5.48)	1.94 (4.29)	2.37 (3.44)	3.00 (4.29)
40-44	3.00 (14.84)	7.68 (19.73)	10.68 (18.54)	6.32 (11.32)	11.34 (16.29)
≥ 45	0.00	0.00 (40.00)	58.82	0.00	0.00

Infants with Down syndrome often have associated anomalies. ACASS does not code minor anomalies associated with Down syndrome, such as single palmar crease, upslanting palpebral fissures, and increased space between the great and second toes. However, most other malformations, if mentioned on the ascertainment documents, are entered routinely into the database. Anomalies such as patent ductus arteriosus, undescended testes and lung hypoplasia are not coded if the infant was born prematurely or weighed less than 2,500 grams. This is consistent with our general coding policies. Table 3 illustrates the proportion of live birth and stillbirth cases of Down Syndrome that have had other anomalies reported over the past 10 years. ToPs have not been included in Table 3.

Table 3. Proportion of Down syndrome cases with and without other anomalies, Alberta live births and stillbirths 1992-2001

Year	Total Cases (n)	Down Syndrome Alone* (n)	Heart Defects Only (n)	GI Defects Only (n)	Single Other † (n)	Multiple ‡ (n)
1992	37	0.43 (16)	0.38 (14)	0.11 (4)	0 (0)	0.08 (3)
1993	46	0.59 (27)	0.30 (14)	0 (0)	0.02 (1)	0.09 (4)
1994	44	0.57 (25)	0.41 (18)	0 (0)	0 (0)	0.02 (1)
1995	50	0.36 (18)	0.52 (26)	0 (0)	0.04 (2)	0.08 (4)
1996	32	0.50 (16)	0.35 (11)	0.09 (3)	0.06 (2)	0 (0)
1997	41	0.42 (17)	0.49 (20)	0.02 (1)	0.05 (2)	0.02 (1)
1998	53	0.51 (27)	0.36 (19)	0 (0)	0.04 (2)	0.09 (5)
1999	45	0.40 (18)	0.33 (15)	0.02 (1)	0.05 (2)	0.20 (9)
2000	54	0.48 (26)	0.28 (15)	0.04 (2)	0.04 (2)	0.16 (9)
2001	57	0.54 (31)	0.19 (11)	0.02 (1)	0.04 (2)	0.21 (12)

* no anomalies reported other than Down syndrome

† Down syndrome with a single anomaly other than heart or GI

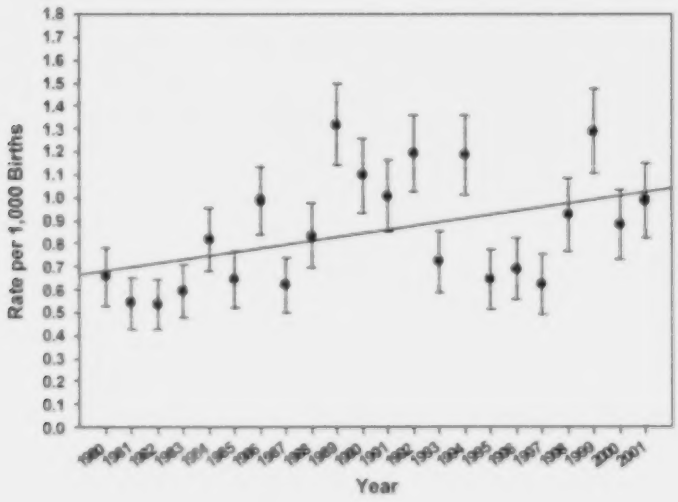
‡ Could include heart or GI defects if other anomalies also reported with the cases

The heart anomalies category, includes only heart defects (no other anomalies) and is comprised mainly of ventriculoseptal defects, atrioseptal defects and endocardial cushion defects. The GI defects include for the most part, duodenal atresia, anorectal stenosis/atresia and tracheo-oesophageal fistula. Again, as with the heart defects, only GI defects are included in this category. The "multiple" category includes cases with Down syndrome who have two or more other anomalies, mainly urinary tract anomalies, hypothyroidism and musculo-skeletal anomalies. Heart defects and GI anomalies might also be included here if reported in association with the other anomalies mentioned above. They would therefore not be counted in the heart and GI defect categories which were reserved for heart and GI defects alone. Few cases reported an isolated anomaly other than a heart or GI abnormality, but those that did included hydronephrosis, hypospadias, hypothyroidism, hydrocephalus and umbilical hernia to name a few.

Limb Reductions

Limb reduction defects are on the rise and will be the focus of more in-depth scrutiny over the coming months. There are a number of possible explanations, aside from a true increase in the rates. Due to computer limitations in the years 1980 – 1996 we were able to code only six anomalies per case. Since 1997, we have been able to code an unlimited number of defects per case. The cases will be reviewed to try to determine whether the upward trend is due to increased ascertainment, different coding procedures or whether the rate has truly risen.

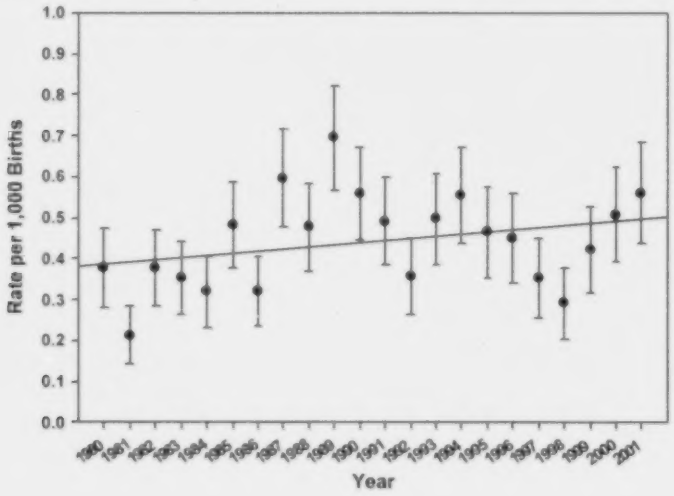
Figure 10. Rate per 1,000 births of limb reduction defects in Alberta, 1980 to 2001



Anorectal atresia/ stenosis

Because differences were noted between ACASS and CCASS (Canadian Congenital Anomalies Surveillance System) data, with respect to anorectal atresia/stenosis, a detailed review is currently underway. Preliminary results indicate that a substantial number of cases belong in the multiple congenital anomaly category VATER/VACTERL.

Figure 11. Rate per 1,000 births of anorectal atresia/stenosis in Alberta, 1980 to 2001.



Anophthalmia/ Microphthalmia

The Alberta rate of anophthalmia/microphthalmia (A/M) in 1999 was noted to be high (2.89/10,000 total births) as was the Canadian Congenital Anomalies Surveillance System (CCASS) rate for that year (1.87/10,000 total births). Most provincial rates for A/M for the years 1991-1999 were approximately 1/10,000 as were the Alberta and CCASS rates (1.3 and 1.2 respectively). ACASS rates from 1980-1998 showed variable rates from 0.3 to 2.1 with the 19-year aggregate being 1.4/10,000. Alberta cases from 1991-2001 were reviewed by a detailed study of each case as reported to ACASS, as well as any accompanying attachments such as ophthalmology and other consultant reports, autopsies, chromosome reports etc. It was not possible to review CCASS cases.

There is considerable etiologic heterogeneity in A/M and it is often difficult to distinguish between them. It is also evident that the etiology of A/M may be the same since an affected person may have anophthalmia in one eye and microphthalmia in the other. Optic fissure closure defects (e.g. coloboma) are also part of the A/M phenotype.

We divided the clinical phenotypes into six categories seen in Table 4. Of the 60 cases, 20 had a chromosome etiology (16 – Trisomy 13, one each with Trisomy 18, Triploidy, 6q- and a 3 deletion/inversion) and 13 had a recognised syndrome or association (Table 5). Sixteen cases had extraocular malformations and four of these had holoprosencephaly (three of four with normal chromosomes). In the 11 years there were only six cases of A/M plus coloboma. There were five other microphthalmia cases with other eye anomalies; two with cataract, two with primary hyperplastic primary vitreous and one with aniridia and corneal opacity, giving a total of 11 cases with ocular defects corresponding to a rate of 0.2/10,000 over the 11-year period 1991-2001. The annual rates are shown in Table 6. Our rate of 1.4/10,000 is very comparable to other registries.

Five cases of Trisomy 13 were reported in 1999. Of note, terminations of pregnancy (ToPs) were not included because ACASS has only been able to ascertain ToP cases from 1997. Since that time four additional A/M cases were ascertained as a result of terminations (two due to Trisomy 13 and two with extra ocular malformations, one of whom had normal chromosomes while the other was a possible amniotic band syndrome. A/M in association with Trisomy 13 may be under ascertained in earlier years.

The inclusion or exclusion of chromosome disorders will make a huge difference to the rates since A/M is a component of at least 40 per cent of Trisomy 13 cases. There are environmental causes of A/M such as rubella, cytomegalic inclusion disease and toxoplasmosis, but we had no such cases in our system. In the past there was a concern that a pesticide (Benomyl) might be responsible for A/M, but studies in England, Italy and Norway showed no support for this.

In conclusion, our review confirmed that the 1999 rate for A/M was high, but was mainly due to five cases of trisomy 13 plus one syndrome case (Meckel-Gruber), leaving three microphthalmia cases. The rates in 2000 were also high (2.4/10,000) and included four chromosomally abnormal cases (three Trisomy 13, one trisomy 18 and two syndrome cases Aicardi and Walker-Warburg and 1 holoprosencephaly) leaving three microphthalmia cases and one anophthalmia case. In 2001 the rate reverted to 1.30/10,000. The review shows how important it is to be able to get back to each individual case.

Table 4. Anophthalmia/Microphthalmia (A/M) Alberta 1991-2001 (Rates per 10 000 total births)

	A/M + coloboma	Eye PLUS	Eye MCA	Chromosome	Syndrome	Total	Rates (95%CI)
1991	-	1	3	2	-	6	1.4 (0.5-3.0)
1992	2	1	1	3	1	8	1.9 (0.8-3.7)
1993	1	-	4	-	3	8	2.0 (0.9-3.9)
1994	1	-	-	-	-	1	0.3 (0.0-1.3)
1995	-	-	-	2	1	3	0.8 (0.2-2.2)
1996	-	-	-	1	-	1	0.3 (0.0-1.3)
1997	-	1	1	2	2	6	1.6 (0.6-3.5)
1998	-	-	2	-	2	4	1.1 (0.3-2.7)
1999	1	2	-	5	1	9	2.4 (1.1-4.5)
2000	1	-	3	3	2	9	2.4 (1.1-4.6)
2001	-	-	2	2	1	5	1.3 (0.4-3.1)
Total	6	5	16	20	13	60	

Eye PLUS = A/M + cataract or aniridia/persistent yperplastic primary vitreous
Eye MCA = A/M + multiple congenital anomalies (includes holoprosencephaly)

Table 5. Syndromes and Associations with Microphthalmia Alberta 1991-2001

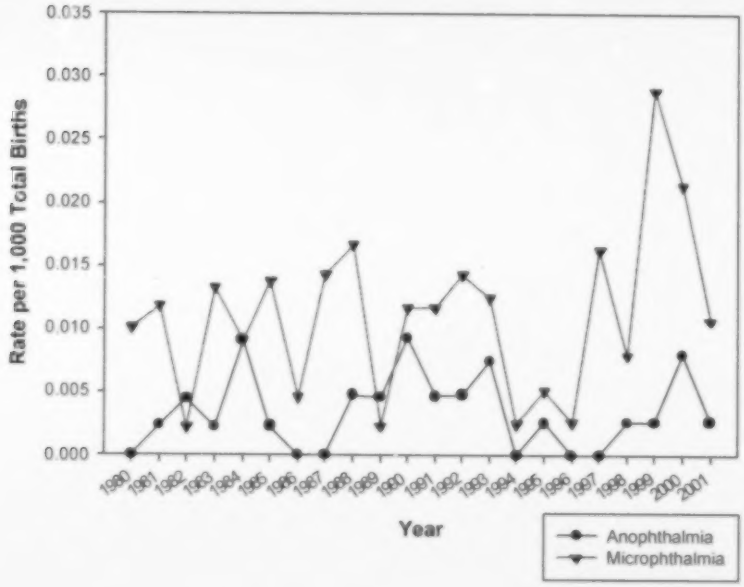
Amniotic Band
CHARGE
Linear Sebaceous
Cohen-Gorlin
Lenz
Nager
Hallermann-Streiff
Complex 1 deficiency
VATER
Meckel-Gruber
Aicardi
Walker-Warburg
Ichthyosis

Table 6. Alberta birth prevalence rates (live and stillbirths) of all cases, isolated cases and Cases with associated anomalies (10,000 total births)

	Number of cases for combined years 1991-2001	Rate per 10,000 total births
All reported cases (isolated and with associated anomalies)	60	1.4
Isolated cases and cases with other eye anomalies combined	11	0.2
Cases associated with HPE, MCA, chromosome anomalies, syndromes	49	1.2

1991-2001 Total births (live + still): 427,960
L = live births S = stillbirths HPE = holoprosencephaly MCA = multiple congenital anomalies

Figure 12. Rate per 1,000 births of anophthalmia/microphthalmia in Alberta, 1980 to 2001



SUMMARY

ACASS reviews anomalies that have been entered into the database on a regular basis. As is evident from the review of anophthalmia and microphthalmia, a detailed study of some individual items helps in the assessment and maintenance of the data quality. With intensive review, some cases might be re-assigned, re-coded or discarded altogether from the database. This continuing review might explain some discepancies in the data from early reports.

Over the past few years, ACASS has been coding anomalies using both the ICD-9/BPA and the Royal College of Paediatrics and Child Health version of ICD-10 coding systems. ACASS has now adopted the ICD-10 classification system and future reports will reflect this change in coding. Earlier data will continue to be stored using the ICD-9/BPA coding scheme. The use of multiple diagnostic coding standards will create challenges for the interpretation of congenital anomaly rates over time. However, ACASS staff are well-positioned to assist in such interpretations.

Additional work is still needed to create meaningful geographic information. The change from 17 regional health authorities to nine means ACASS will need to develop alternative approaches to the geographic analysis of congenital anomaly rates to ensure areas with perceived high rates can be investigated.

APPENDICES

Appendix A1	**Section XIV (740.00 - 759.99), anomaly rates per 1,000 total births**
Appendix A2	**Selected anomalies with rates of live births and stillbirths compared with total rates including terminations of pregnancy (ToP)**
Appendix A3	**Numbers of cases, anomalies and anomalies per case 1980-2001**
Appendix A4	**Termination of pregnancy (ToP) not registered with Alberta Vital Statistics (gestation <20 weeks or birth weight <500g)**

Appendix A1. Section XIV (740.00 - 759.99), Anomaly Rates per 1,000 Total Births

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
ANENCEPHALY AND SIMILAR ANOMALIES (740)	**NUMBER**	**147**	**76**	**6**	**8**	**3**
	RATE	**0.34**	**0.19**	**0.16**	**0.22**	**0.08**
	Lower CI	**0.29**	**0.15**	**0.06**	**0.09**	**0.02**
	Upper CI	**0.40**	**0.24**	**0.34**	**0.42**	**0.23**
Anencephaly (7400)	NUMBER	142	66	4	8	3
	RATE	0.33	0.17	0.11	0.22	0.08
	Lower CI	0.28	0.13	0.03	0.09	0.02
	Upper CI	0.39	0.21	0.26	0.42	0.23
Craniorachischisis (7401)	NUMBER	4	10	2	0	0
	RATE	0.01	0.03	0.05		
	Lower CI	0.00	0.01	0.01		
	Upper CI	0.02	0.05	0.18		
Iniencephaly (7402)	NUMBER	1	0	0	0	0
	RATE	0.00				
	Lower C	0.00				
	Upper CI	0.01				
SPINA BIFIDA (741)	**NUMBER**	**219**	**179**	**13**	**7**	**11**
	RATE	**0.51**	**0.45**	**0.34**	**0.19**	**0.29**
	Lower CI	**0.44**	**0.39**	**0.18**	**0.08**	**0.15**
	Upper CI	**0.58**	**0.52**	**0.58**	**0.39**	**0.52**
with Hydrocephaly (7410)	NUMBER	135	95	6	4	3
	RATE	0.31	0.24	0.16	0.11	0.08
	Lower CI	0.26	0.19	0.06	0.03	0.02
	Upper CI	0.37	0.29	0.34	0.27	0.23
without Hydrocephaly (7419)	NUMBER	84	84	7	3	8
	RATE	0.20	0.21	0.18	0.08	0.21
	Lower CI	0.16	0.17	0.07	0.02	0.09
	Upper CI	0.24	0.26	0.38	0.23	0.42

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
OTHER CONGENITAL ANOMALIES OF NERVOUS SYSTEM (742)	**NUMBER**	**682**	**670**	**70**	**87**	**102**
	RATE	**1.58**	**1.69**	**1.84**	**2.36**	**2.72**
	Lower CI	**1.47**	**1.56**	**1.44**	**1.89**	**2.22**
	Upper CI	**1.71**	**1.82**	**2.32**	**2.91**	**3.30**
Encephalocele (7420)	NUMBER	41	33	2	4	6
	RATE	0.10	0.08	0.05	0.11	0.16
	Lower CI	0.07	0.06	0.01	0.03	0.06
	Upper CI	0.13	0.12	0.18	0.27	0.34
Microcephaly (7421)	NUMBER	149	110	11	14	15
	RATE	0.35	0.28	0.29	0.38	0.40
	Lower CI	0.29	0.23	0.14	0.21	0.22
	Upper CI	0.41	0.33	0.52	0.64	0.66
Reduction Deformities of Brain (7422)	NUMBER	113	169	17	17	25
	RATE	0.26	0.43	0.45	0.46	0.67
	Lower CI	0.22	0.36	0.26	0.27	0.43
	Upper CI	0.32	0.50	0.71	0.74	0.98
Congenital Hydrocephaly (7423)	NUMBER	236	190	19	27	21
	RATE	0.55	0.48	0.50	0.73	0.56
	Lower CI	0.48	0.41	0.30	0.48	0.35
	Upper CI	0.62	0.55	0.78	1.07	0.86
Other Specified Anomalies of Brain (7424)	NUMBER	123	150	19	18	27
	RATE	0.29	0.38	0.50	0.79	0.72
	Lower CI	0.24	0.32	0.30	0.29	0.48
	Upper CI	0.34	0.44	0.78	0.77	1.05
Other Specified Anomalies of Spinal Cord (7425)	NUMBER	6	17	2	6	7
	RATE	0.01	0.04	0.05	0.16	0.19
	Lower CI	0.01	0.03	0.01	0.06	0.08
	Upper CI	0.03	0.07	0.18	0.35	0.38
Other Specified Anomalies of Nervous System (7428)	NUMBER	9	4	0	0	1
	RATE	0.02	0.01			0.03
	Lower CI	0.01	0.00			0.00
	Upper CI	0.04	0.03			0.13

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Unspecified Anomalies of Brain, Spinal Cord and Nervous System (7429)	NUMBER	5	2	0	1	0
	RATE	0.01	0.01		0.03	
	Low er CI	0.00	0.00		0.00	
	Upper CI	0.03	0.02		0.14	
CONGENITAL ANOMALIES OF EYE (743)	**NUMBER**	**678**	**471**	**51**	**30**	**32**
	RATE	**1.57**	**1.19**	**1.34**	**0.81**	**0.85**
	Lower CI	**1.46**	**1.08**	**1.00**	**0.55**	**0.58**
	Upper CI	**1.70**	**1.30**	**1.76**	**1.16**	**1.21**
Anophthalmos (7430)	NUMBER	13	13	0	3	2
	RATE	0.03	0.03		0.08	0.05
	Lower CI	0.02	0.02		0.02	0.01
	Upper CI	0.05	0.06		0.23	0.18
Microphthalmos (7431)	NUMBER	42	45	11	8	4
	RATE	0.10	0.11	0.29	0.22	0.11
	Lower CI	0.07	0.08	0.14	0.09	0.03
	Upper CI	0.13	0.15	0.52	0.42	0.27
Buphthalmos (7432)	NUMBER	14	10	1	1	0
	RATE	0.03	0.03	0.03	0.03	
	Lower CI	0.02	0.01	0.00	0.00	
	Upper CI	0.05	0.05	0.13	0.14	
Cataract and Lens Anomalies (7433)	NUMBER	62	58	9	3	3
	RATE	0.14	0.15	0.24	0.08	0.08
	Lower CI	0.11	0.11	0.11	0.02	0.02
	Upper CI	0.18	0.19	0.45	0.23	0.23
Coloboma and Other Anomalies of Anterior Segments (7434)	NUMBER	51	52	11	1	4
	RATE	0.12	0.13	0.29	0.03	0.11
	Lower CI	0.09	0.10	0.14	0.00	0.03
	Upper CI	0.16	0.17	0.52	0.14	0.27
Anomalies of Posterior Segments (7435)	NUMBER	27	30	6	3	4
	RATE	0.06	0.08	0.16	0.08	0.11
	Lower CI	0.04	0.05	0.06	0.02	0.03
	Upper CI	0.09	0.11	0.34	0.23	0.27

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Anomalies of Eyelids, Lacrimal System and Orbit (7436)	NUMBER	436	242	11	10	13
	RATE	1.01	0.61	0.29	0.27	0.35
	Lower CI	0.92	0.54	0.14	0.13	0.18
	Upper CI	1.11	0.69	0.52	0.50	0.59
Other Specified Anomalies of Eye (7438)	NUMBER	28	20	2	1	2
	RATE	0.07	0.05	0.05	0.03	0.05
	Lower CI	0.04	0.03	0.01	0.00	0.01
	Upper CI	0.09	0.08	0.18	0.14	0.18
Unspecified Anomalies of Eye (7439)	NUMBER	5	1	0	0	0
	RATE	0.01	0.00			
	Lower CI	0.00	0.00			
	Upper CI	0.03	0.01			
CONGENITAL ANOMALIES OF EAR, FACE AND NECK (744)	**NUMBER**	**1193**	**1134**	**114**	**90**	**124**
	RATE	**2.77**	**2.86**	**3.00**	**2.44**	**3.31**
	Lower CI	**2.62**	**2.70**	**2.47**	**1.97**	**2.75**
	Upper CI	**2.93**	**3.03**	**3.60**	**3.00**	**3.94**
Anomalies of Ear Causing Hearing Impairment (7440)	NUMBER	44	57	13	8	5
	RATE	1.10	0.14	0.34	0.22	0.13
	Lower CI	0.07	0.11	0.18	0.09	0.04
	Upper CI	0.14	0.19	0.58	0.42	0.31
Accessory Auricle (7441)	NUMBER	789	752	65	61	84
	RATE	1.83	1.90	1.71	1.66	2.24
	Lower CI	1.71	1.76	1.32	1.27	1.79
	Upper CI	1.96	2.04	2.18	2.13	2.77
Other Specified Anomalies of Ear (7442)	NUMBER	134	153	15	9	19
	RATE	0.31	0.39	0.39	0.24	0.51
	Lower CI	0.26	0.33	0.22	0.11	0.31
	Upper CI	0.37	0.45	0.65	0.46	0.79
Unspecified Anomalies of Ear (7443)	NUMBER	74	31	5	2	0
	RATE	0.17	0.08	0.13	0.05	
	Lower CI	0.14	0.05	0.04	0.01	
	Upper CI	0.22	0.11	0.30	0.19	

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Branchial Cleft, Cyst or Fistula; Preauricular Sinus (7444)	NUMBER	117	113	8	7	13
	RATE	0.27	0.28	0.21	0.19	0.35
	Lower CI	0.22	0.23	0.09	0.08	0.18
	Upper CI	0.33	0.34	0.41	0.39	0.59
Webbing of Neck (7445)	NUMBER	11	10	4	2	1
	RATE	0.03	0.03	0.11	0.05	0.03
	Lower CI	0.01	0.01	0.03	0.01	0.00
	Upper CI	0.05	0.05	0.26	0.19	0.13
Other Specified Anomalies of Face and Neck (7448)	NUMBER	17	18	4	1	2
	RATE	0.04	0.05	0.11	0.03	0.05
	Lower CI	0.02	0.03	0.03	0.00	0.01
	Upper CI	0.06	0.07	0.26	0.13	0.18
Unspecified Anomalies of Face and Neck (7449)	NUMBER	7	0	0	0	0
	RATE	0.02				
	Lower CI	0.01				
	Upper CI	0.03				
BULBUS CORDIS ANOMALIES AND ANOMALIES OF CARDIAC SEPTAL CLOSURE (745)	**NUMBER**	**2226**	**2382**	**197**	**195**	**211**
	RATE	**5.17**	**6.01**	**5.18**	**5.29**	**5.63**
	Lower CI	**4.96**	**5.77**	**4.48**	**4.58**	**4.90**
	Upper CI	**5.39**	**6.25**	**5.96**	**6.09**	**6.44**
Common Truncus (7450)	NUMBER	39	30	2	3	1
	RATE	0.09	0.08	0.05	0.08	0.03
	Lower CI	0.06	0.05	0.01	0.02	0.00
	Upper CI	0.12	0.11	0.18	0.23	0.13
Transposition of Great Vessels (7451)	NUMBER	127	125	12	7	18
	RATE	0.29	0.32	0.32	0.19	0.48
	Lower CI	0.25	0.26	0.16	0.08	0.29
	Upper CI	0.35	0.38	0.55	0.39	0.76
Tetralogy of Fallot (7452)	NUMBER	99	115	12	14	9
	RATE	0.23	0.29	0.32	0.38	0.24
	Lower CI	0.19	0.24	0.16	0.21	0.11
	Upper CI	0.28	0.35	0.55	0.64	0.45

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Common Ventricle (7453)	NUMBER	42	79	10	7	6
	RATE	0.10	0.20	0.26	0.19	0.16
	Lower CI	0.07	0.16	0.13	0.08	0.04
	Upper CI	0.13	0.25	0.48	0.39	0.34
Ventricular Septal Defect (7454)	NUMBER	1208	1121	86	98	99
	RATE	2.81	2.83	2.26	2.66	2.64
	Lower CI	2.65	2.66	1.81	2.16	2.15
	Upper CI	2.97	3.00	2.79	3.24	3.21
Ostium Secundum Type Atrial Septal Defect (7455)	NUMBER	561	761	53	50	55
	RATE	1.30	1.92	1.39	1.36	1.47
	Lower CI	1.20	1.79	1.04	1.01	1.11
	Upper CI	1.42	2.06	1.82	1.79	1.91
Endocardial Cushion Defects (7456)	NUMBER	135	150	22	16	23
	RATE	0.31	0.38	0.58	0.43	0.61
	Lower CI	0.26	0.32	0.36	0.25	0.39
	Upper CI	0.37	0.44	0.88	0.70	0.92
Cor Biloculare (7457)	NUMBER	2	1	0	0	0
	RATE	0.00	0.00			
	Lower CI	0.00	0.00			
	Upper CI	0.02	0.01			
Other (7458)	NUMBER	5	0	0	0	0
	RATE	0.01				
	Lower CI	0.00				
	Upper CI	0.03				
Unspecified Defect of Septal Closure (7459)	NUMBER	5	0	0	0	0
	RATE	0.01				
	Lower CI	0.00				
	Upper CI	0.03				
OTHER CONGENITAL ANOMALIES OF THE HEART (746)	**NUMBER**	**986**	**1043**	**98**	**80**	**91**
	RATE	**2.29**	**2.63**	**2.58**	**2.17**	**2.43**
	Lower CI	**2.15**	**2.47**	**2.09**	**1.72**	**1.96**
	Upper CI	**2.44**	**2.80**	**3.14**	**2.70**	**2.98**

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Anomalies of Pulmonary Valve (7460)	NUMBER	346	284	23	21	29
	RATE	0.80	0.72	0.60	0.57	0.77
	Lower CI	0.72	0.64	0.38	0.35	0.52
	Upper CI	0.89	0.80	0.91	0.87	1.11
Tricuspid Atresia and Stenosis (7461)	NUMBER	61	117	9	2	5
	RATE	0.14	0.30	0.24	0.05	0.13
	Lower CI	0.11	0.24	0.11	0.01	0.04
	Upper CI	0.18	0.35	0.45	0.19	0.31
Ebstein's Anomaly (7462)	NUMBER	20	20	5	3	2
	RATE	0.05	0.05	0.13	0.08	0.05
	Lower CI	0.03	0.03	0.04	0.02	0.01
	Upper CI	0.07	0.08	0.30	0.23	0.18
Stenosis of Aortic Valve (7463)	NUMBER	53	70	6	10	6
	RATE	0.12	0.18	0.16	0.27	0.16
	Lower CI	0.09	0.14	0.06	0.13	0.06
	Upper CI	0.16	0.22	0.34	0.50	0.34
Insufficiency of Aortic Valve (7464)	NUMBER	48	87	9	8	13
	RATE	0.11	0.22	0.24	0.22	0.35
	Lower CI	0.08	0.18	0.11	0.09	0.18
	Upper CI	0.15	0.27	0.45	0.42	0.59
Mitral Stenosis (7465)	NUMBER	28	25	4	4	1
	RATE	0.07	0.06	0.11	0.11	0.03
	Lower CI	0.04	0.04	0.03	0.03	0.00
	Upper CI	0.09	0.09	0.26	0.27	0.13
Mitral Insufficiency (7466)	NUMBER	16	75	10	5	6
	RATE	0.04	0.19	0.26	0.14	0.16
	Lower CI	0.02	0.15	0.13	0.04	0.06
	Upper CI	0.06	0.24	0.48	0.31	0.34
Hypoplastic Left Heart Syndrome (7467)	NUMBER	126	129	12	16	14
	RATE	0.29	0.33	0.32	0.43	0.37
	Lower CI	0.24	0.27	0.16	0.25	0.20
	Upper CI	0.35	0.39	0.55	0.70	0.63

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Other Specified Anomalies of Heart (7468)	NUMBER	219	228	20	11	15
	RATE	0.51	0.57	0.53	0.30	0.40
	Lower CI	0.44	0.50	0.32	0.15	0.22
	Upper CI	0.58	0.65	0.81	0.53	0.66
Unspecified Anomalies of Heart (7469)	NUMBER	69	6	0	0	0
	RATE	0.16	0.02			
	Lower CI	0.12	0.01			
	Upper CI	0.20	0.03			
OTHER CONGENITAL ANOMALIES OF CIRCULATORY SYSTEM (747)	**NUMBER**	**1804**	**1120**	**102**	**97**	**85**
	RATE	**4.19**	**2.82**	**2.68**	**2.63**	**2.27**
	Lower CI	**4.00**	**2.66**	**2.19**	**2.14**	**1.81**
	Upper CI	**4.39**	**3.00**	**3.26**	**3.21**	**2.80**
Patent Ductus Arteriosus (7470)	NUMBER	1057	320	22	24	18
	RATE	2045	0.81	0.58	0.65	0.48
	Lower CI	2.31	0.72	0.36	0.42	0.29
	Upper CI	2.61	0.90	0.87	0.97	0.76
Coarctation of the Aorta (7471)	NUMBER	172	196	15	15	13
	RATE	0.40	0.49	0.39	0.41	0.35
	Lower CI	0.34	0.43	0.22	0.23	0.18
	Upper CI	0.46	0.57	0.65	0.67	0.59
Anomalies of Aorta (7472)	NUMBER	60	52	7	6	5
	RATE	0.14	0.13	0.18	0.16	0.13
	Lower CI	0.11	0.10	0.07	0.06	0.04
	Upper CI	0.18	0.17	0.38	0.35	0.31
Anomalies of Pulmonary Artery (7473)	NUMBER	97	127	11	9	6
	RATE	0.23	0.32	0.29	0.24	0.16
	Lower CI	0.18	0.27	0.14	0.11	0.06
	Upper CI	0.27	0.38	0.52	0.46	0.34
Anomalies of Great Veins (7474)	NUMBER	80	96	19	16	9
	RATE	0.19	0.24	0.50	0.43	0.24
	Lower CI	0.15	0.20	0.30	0.25	0.11
	Upper CI	0.23	0.30	0.78	0.70	0.45

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Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Absence or Hypoplasia of Umbilical Artery (7475)	NUMBER	144	185	21	18	26
	RATE	0.33	0.47	0.55	0.49	0.69
	Lower CI	0.28	0.40	0.34	0.29	0.45
	Upper CI	0.39	0.54	0.84	0.77	1.02
Other Anomalies of Peripheral Vascular System (7476)	NUMBER	23	45	5	4	6
	RATE	0.05	0.11	0.13	0.11	0.16
	Lower CI	0.03	0.08	0.04	0.03	0.06
	Upper CI	0.08	0.15	0.30	0.27	0.34
Other Specified Anomalies of Circulatory System (7478)	NUMBER	168	98	2	5	2
	RATE	0.39	0.25	0.05	0.14	0.05
	Lower CI	0.33	0.20	0.01	0.04	0.01
	Upper CI	0.45	0.30	0.18	0.31	0.18
Unspecified Anomalies of Circulatory System (7479)	NUMBER	3	0	0	0	0
	RATE	0.01				
	Lower CI	0.00				
	Upper CI	0.02				
CONGENITAL ANOMALIES OF RESPIRATORY SYSTEM (748)	**NUMBER**	**317**	**200**	**17**	**23**	**20**
	RATE	**0.74**	**0.50**	**0.45**	**0.62**	**0.53**
	Lower CI	**0.66**	**0.44**	**0.26**	**0.40**	**0.33**
	Upper CI	**0.82**	**0.58**	**0.71**	**0.94**	**0.82**
Choanal Atresia (7480)	NUMBER	56	51	1	2	5
	RATE	0.13	0.13	0.03	0.05	0.13
	Lower CI	0.10	0.10	0.00	0.01	0.04
	Upper CI	0.17	0.17	0.13	0.19	0.31
Other Anomalies of Nose (7481)	NUMBER	21	16	1	1	1
	RATE	0.05	0.04	0.03	0.03	0.03
	Lower CI	0.03	0.02	0.00	0.00	0.00
	Upper CI	0.07	0.07	0.13	0.14	0.13
Web of Larynx (7482)	NUMBER	9	3	0	0	1
	RATE	0.02	0.01			0.03
	Lower CI	0.01	0.00			0.00
	Upper CI	0.04	0.02			0.13

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Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Other Anomalies of Larynx, Trachea and Bronchus (7483)	NUMBER	40	39	2	6	4
	RATE	0.09	0.10	0.05	0.16	0.11
	Lower CI	0.07	0.07	0.01	0.06	0.03
	Upper CI	0.13	0.13	0.18	0.35	0.27
Cystic Lung (7484)	NUMBER	9	20	1	5	4
	RATE	0.02	0.05	0.03	0.14	0.11
	Lower CI	0.01	0.03	0.00	0.04	0.03
	Upper CI	0.04	0.08	0.13	0.31	0.27
Agenesis, Hypoplasia and Dysplasia of Lung (7485)	NUMBER	167	54	9	6	4
	RATE	0.39	0.14	0.24	0.16	0.11
	Lower CI	0.33	0.10	0.11	0.06	0.03
	Upper CI	0.45	0.18	0.45	0.35	0.27
Other Anomalies of Lung (7486)	NUMBER	13	12	3	1	1
	RATE	0.03	0.03	0.08	0.03	0.03
	Lower CI	0.02	0.02	0.02	0.00	0.00
	Upper CI	0.05	0.05	0.22	0.14	0.13
Other Specified Anomalies of Respiratory System (7488)	NUMBER	2	5	0	2	0
	RATE	0.00	0.01		0.05	
	Lower CI	0.00	0.00		0.01	
	Upper CI	0.02	0.03		0.19	
CLEFT PALATE AND CLEFT LIP (749)	**NUMBER**	**767**	**789**	**71**	**70**	**79**
	RATE	**1.78**	**1.99**	**1.87**	**1.90**	**2.11**
	Lower CI	**1.66**	**1.85**	**1.46**	**1.48**	**1.67**
	Upper CI	**1.91**	**2.13**	**2.35**	**2.40**	**2.63**
Cleft Palate (7490)	NUMBER	291	329	37	30	34
	RATE	0.68	0.83	0.97	0.81	0.91
	Lower CI	0.60	0.74	0.69	0.55	0.63
	Upper CI	0.76	0.92	1.34	1.16	1.27
Cleft Lip (7491)	NUMBER	144	173	9	18	11
	RATE	0.33	0.44	0.24	0.49	0.29
	Lower CI	0.28	0.37	0.11	0.29	0.15
	Upper CI	0.39	0.51	0.45	0.77	0.52

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Cleft Palate with Cleft Lip (7492)	NUMBER	332	287	25	22	34
	RATE	0.77	0.72	0.66	0.60	0.91
	Lower CI	0.69	0.64	0.43	0.37	0.63
	Upper CI	0.86	0.81	0.97	0.90	1.27
OTHER CONGENITAL ANOMALIES OF UPPER ALIMENTARY TRACT (750)	**NUMBER**	**588**	**436**	**45**	**43**	**47**
	RATE	**1.37**	**1.10**	**1.18**	**1.17**	**1.25**
	Lower CI	**1.26**	**1.00**	**0.86**	**0.85**	**0.92**
	Upper CI	**1.48**	**1.21**	**1.58**	**1.57**	**1.67**
Other Anomalies of Tongue (7501)	NUMBER	30	23	1	1	2
	RATE	0.07	0.06	0.03	0.03	0.05
	Lower CI	0.05	0.04	0.00	0.00	0.01
	Upper CI	0.10	0.09	0.13	0.14	0.18
Other Specified Anomalies of Mouth and Pharynx (7502)	NUMBER	22	29	4	0	1
	RATE	0.05	0.07	0.11		0.03
	Lower CI	0.03	0.05	0.03		0.00
	Upper CI	0.08	0.11	0.26		0.13
Tracheo-Oesophageal Fistula, Oesophageal Atresia & Stenosis (7503)	NUMBER	128	92	13	5	6
	RATE	0.30	0.23	0.34	0.14	0.16
	Lower CI	0.25	0.19	0.18	0.04	0.06
	Upper CI	0.35	0.28	0.58	0.31	0.34
Other Specified Anomalies of Oesophagus (7504)	NUMBER	2	1	0	0	2
	RATE	0.00	0.00			0.05
	Lower CI	0.00	0.00			0.01
	Upper CI	0.02	0.01			0.18
Hypertrophic Pyloric Stenosis (7505)	NUMBER	395	281	25	33	35
	RATE	0.92	0.71	0.66	0.90	0.93
	Lower CI	0.83	0.63	0.43	0.62	0.65
	Upper CI	1.01	0.80	0.97	1.26	1.30
Hiatus Hernia (7506)	NUMBER	5	6	2	2	1
	RATE	0.01	0.02	0.05	0.05	0.03
	Lower CI	0.00	0.01	0.01	0.01	0.00
	Upper CI	0.03	0.03	0.18	0.19	0.13

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Other Specified Anomalies of Stomach (7507)	NUMBER	3	1	0	2	0
	RATE	0.01	0.00		0.05	
	Lower CI	0.00	0.00		0.01	
	Upper CI	0.02	0.01		0.19	
Unspecified Anomalies of Upper Alimentary Tract (7509)	NUMBER	1	0	0	0	0
	RATE	0.00				
	Lower CI	0.00				
	Upper CI	0.01				
OTHER CONGENITAL ANOMALIES OF THE DIGESTIVE SYSTEM (751)	**NUMBER**	**631**	**746**	**104**	**76**	**74**
	RATE	**1.47**	**1.88**	**2.73**	**2.06**	**1.97**
	Lower CI	**1.35**	**1.75**	**2.24**	**1.63**	**1.55**
	Upper CI	**1.58**	**2.02**	**3.31**	**2.58**	**2.48**
Meckel's Diverticulum (7510)	NUMBER	48	38	5	4	5
	RATE	0.11	0.10	0.13	0.11	0.13
	Lower CI	0.08	0.07	0.04	0.03	0.04
	Upper CI	0.15	0.13	0.30	0.27	0.31
Atresia and Stenosis of Small Intestine (7511)	NUMBER	95	119	21	15	12
	RATE	0.22	0.30	0.55	0.41	0.32
	Lower CI	0.18	0.25	0.34	0.23	0.17
	Upper CI	0.27	0.36	0.84	0.67	0.56
Atresia and Stenosis of Large Intestine, Rectum and Anal Canal (7512)	NUMBER	197	204	23	21	20
	RATE	0.46	0.51	0.60	0.57	0.53
	Lower CI	0.40	0.45	0.38	0.35	0.33
	Upper CI	0.53	0.59	0.91	0.87	0.82
Hirschsprung's Disease and Other Functional Disorders of Colon (7513)	NUMBER	63	55	6	4	3
	RATE	0.15	0.14	0.16	0.11	0.08
	Lower CI	0.11	0.10	0.06	0.03	0.02
	Upper CI	0.19	0.18	0.34	0.27	0.23
Anomalies of Intestinal Fixation (7514)	NUMBER	111	149	23	11	16
	RATE	0.26	0.38	0.60	0.30	0.43
	Lower CI	0.21	0.32	0.38	0.15	0.24
	Upper CI	0.31	0.44	0.91	0.53	0.69

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Other Anomalies of Intestine (7515)	NUMBER	64	109	21	14	11
	RATE	0.15	0.27	0.55	0.38	0.29
	Lower CI	0.11	0.23	0.34	0.21	0.15
	Upper CI	0.19	0.33	0.84	0.64	0.52
Anomalies of Gall Bladder, Bile Ducts and Liver (7516)	NUMBER	41	52	2	6	4
	RATE	0.10	0.13	0.05	0.16	0.11
	Lower CI	0.07	0.10	0.01	0.06	0.03
	Upper CI	0.13	0.17	0.18	0.35	0.27
Anomalies of Pancreas (7517)	NUMBER	9	20	3	1	3
	RATE	0.02	0.05	0.08	0.03	0.08
	Lower CI	0.01	0.03	0.02	0.00	0.02
	Upper CI	0.04	0.08	0.22	0.14	0.23
Other Specified Anomalies of Digestive System (7518)	NUMBER	1	0	0	0	0
	RATE	0.00				
	Lower CI	0.00				
	Upper CI	0.01				
Unspecified Anomalies of Digestive System (7519)	NUMBER	2	0	0	0	0
	RATE	0.00				
	Lower CI	0.00				
	Upper CI	0.02				
CONGENITAL ANOMALIES OF GENITAL ORGANS (752)	**NUMBER**	**2473**	**2239**	**182**	**201**	**200**
	RATE	**5.74**	**5.65**	**4.79**	**5.46**	**5.34**
	Lower CI	**5.52**	**5.42**	**4.12**	**4.73**	**4.62**
	Upper CI	**5.97**	**5.89**	**5.53**	**6.26**	**6.13**
Anomalies of Ovaries (7520) *	NUMBER	12	11	1	1	6
	RATE	0.06	0.06	0.05	0.06	0.33
	Lower CI	0.03	0.03	0.00	0.00	0.12
	Upper CI	0.10	0.10	0.27	0.28	0.71
Anomalies of Fallopian Tubes and Broad Ligaments (7521) *	NUMBER	4	1	0	0	0
	RATE	0.02	0.01			
	Lower CI	0.01	0.00			
	Upper CI	0.05	0.03			

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Doubling of Uterus (7522) *	NUMBER	5	1	0	1	0
	RATE	0.02	0.01		0.06	
	Lower CI	0.01	0.00		0.00	
	Upper CI	0.06	0.03		0.28	
Other Anomalies of Uterus (7523) *	NUMBER	14	28	5	3	3
	RATE	0.07	0.14	0.27	0.17	0.16
	Lower CI	0.04	0.10	0.09	0.03	0.03
	Upper CI	0.11	0.21	0.61	0.47	0.47
Anomalies of Cervix, Vagina and External Female Genitalia (7524) *	NUMBER	33	29	8	4	1
	RATE	0.16	0.15	0.43	0.22	0.05
	Lower CI	0.11	0.10	0.19	0.06	0.00
	Upper CI	0.22	0.22	0.84	0.55	0.28
Undescended Testicle (7525) †	NUMBER	1182	1005	75	84	89
	RATE	5.39	4.94	3.88	4.49	4.63
	Lower CI	5.08	4.64	3.05	3.58	3.72
	Upper CI	5.70	5.26	4.86	5.55	5.69
Hypospadias and Epispadias (7526) †	NUMBER	996	925	70	86	80
	RATE	4.54	4.55	3.62	4.59	4.16
	Lower CI	4.26	4.26	2.82	3.68	3.30
	Upper CI	4.83	4.85	4.57	5.67	5.17
Indeterminate Sex and Pseudohermaphroditism (7527)	NUMBER	22	35	5	6	3
	RATE	0.05	0.09	0.13	0.16	0.08
	Lower CI	0.03	0.06	0.04	0.06	0.02
	Upper CI	0.08	0.12	0.30	0.35	0.23
Other Specified Anomalies of Male Genital Organs (7528) †	NUMBER	186	204	18	16	18
	RATE	0.85	1.00	0.93	0.85	0.94
	Lower CI	0.73	0.87	0.55	0.49	0.56
	Upper CI	0.98	1.15	1.47	1.39	1.48
Unspecified Anomalies of Genital Organs (7529)	NUMBER	3	0	0	0	0
	RATE	0.01				
	Lower CI	0.00				
	Upper CI	0.02				

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
CONGENITAL ANOMALIES OF URINARY SYSTEM (753)	**NUMBER**	**864**	**1295**	**162**	**144**	**176**
	RATE	**2.01**	**3.27**	**4.26**	**3.91**	**4.70**
	Lower CI	**1.87**	**3.09**	**3.63**	**3.30**	**4.03**
	Upper CI	**2.14**	**3.45**	**4.97**	**4.60**	**5.44**
Renal Agenesis and Dysgenesis (7530)	NUMBER	165	183	19	17	21
	RATE	0.38	0.46	0.50	0.46	0.56
	Lower CI	0.33	0.40	0.30	0.27	0.35
	Upper CI	0.45	0.53	0.78	0.74	0.86
Cystic Kidney Disease (7531)	NUMBER	130	195	21	23	17
	RATE	0.30	0.49	0.55	0.62	0.45
	Lower CI	0.25	0.43	0.34	0.40	0.26
	Upper CI	0.36	0.57	0.84	0.94	0.73
Obstructive Defects of Renal Pelvis and Ureter (7532)	NUMBER	325	595	76	59	87
	RATE	0.75	1.50	2.00	1.60	2.32
	Lower CI	0.67	1.38	1.58	1.22	1.86
	Upper CI	0.84	1.63	2.50	2.07	2.86
Other Specified Anomalies of Kidney (7533)	NUMBER	70	118	20	24	26
	RATE	0.16	0.30	0.53	0.65	0.69
	Lower CI	0.13	0.25	0.32	0.42	0.45
	Upper CI	0.21	0.36	0.81	0.97	1.02
Other Specified Anomalies of Ureter (7534)	NUMBER	61	88	15	4	12
	RATE	0.14	0.22	0.39	0.11	0.32
	Lower CI	0.11	0.18	0.22	0.03	0.17
	Upper CI	0.18	0.27	0.65	0.27	0.56
Exstrophy of Urinary Bladder (7535)	NUMBER	13	12	0	1	4
	RATE	0.03	0.03		0.03	0.11
	Lower CI	0.02	0.02		0.00	0.03
	Upper CI	0.05	0.05		0.14	0.27
Atresia and Stenosis of Urethra and Bladder Neck (7536)	NUMBER	63	52	6	8	5
	RATE	0.15	0.13	0.16	0.22	0.13
	Low er CI	0.11	0.10	0.06	0.09	0.04
	Upper CI	0.19	0.17	0.34	0.42	0.31

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Anomalies of Urachus (7537)	NUMBER	7	17	1	1	3
	RATE	0.02	0.04	0.03	0.03	0.08
	Lower CI	0.01	0.03	0.00	0.00	0.02
	Upper CI	0.03	0.07	0.13	0.14	0.23
Other Specified Anomalies of Bladder and Urethra (7538)	NUMBER	29	32	4	7	1
	RATE	0.07	0.08	0.11	0.19	0.03
	Lower CI	0.05	0.06	0.03	0.08	0.00
	Upper CI	0.10	0.11	0.26	0.39	0.13
CERTAIN CONGENITAL MUSCULO-SKELETAL DEFORMITIES (754)	**NUMBER**	**3462**	**2422**	**174**	**177**	**172**
	RATE	**8.04**	**6.11**	**4.58**	**4.80**	**4.59**
	Lower CI	**7.77**	**5.87**	**3.92**	**4.12**	**3.93**
	Upper CI	**8.31**	**6.36**	**5.31**	**5.57**	**5.33**
Of Skull, Face and Jaw (7540)	NUMBER	58	39	3	5	6
	RATE	0.13	0.10	0.08	0.14	0.16
	Lower CI	0.10	0.07	0.02	0.04	0.06
	Upper CI	0.17	0.13	0.22	0.31	0.34
Of Sternocleidomastoid Muscle (7541)	NUMBER	13	17	1	0	0
	RATE	0.03	0.04	0.03		
	Lower CI	0.02	0.03	0.00		
	Upper CI	0.05	0.07	0.13		
Of Spine (7542)	NUMBER	23	24	3	4	1
	RATE	0.05	0.06	0.08	0.11	0.03
	Lower CI	0.03	0.04	0.02	0.03	0.00
	Upper CI	0.08	0.09	0.22	0.27	0.13
Congenital Dislocation of Hip (7543)	NUMBER	1164	828	61	74	63
	RATE	2.70	2.09	1.60	2.01	1.68
	Lower CI	2.55	1.95	1.23	1.58	1.29
	Upper CI	2.86	2.24	2.06	2.52	2.15
Genu Recurvatum and Bowing of Long Bones of Leg (7544)	NUMBER	6	22	2	1	0
	RATE	0.01	0.06	0.05	0.03	
	Lower CI	0.01	0.03	0.01	0.00	
	Upper CI	0.03	0.08	0.18	0.14	

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Varus Deformities of Feet (7545)	NUMBER	1335	919	54	54	49
	RATE	3.10	2.32	1.42	1.47	1.31
	Lower CI	2.94	2.17	1.07	1.10	0.97
	Upper CI	3.27	2.47	1.85	1.91	1.73
Valgus Deformities of Feet (7546)	NUMBER	480	256	12	11	13
	RATE	1.11	0.65	0.32	0.30	0.35
	Lower CI	1.02	0.57	0.16	0.15	0.18
	Upper CI	1.22	0.73	0.55	0.53	0.59
Other Deformities of Feet (7547)	NUMBER	314	285	34	27	35
	RATE	0.73	0.72	0.89	0.73	0.93
	Lower CI	0.65	0.64	0.62	0.48	0.65
	Upper CI	0.81	0.81	1.25	1.07	1.30
Other Specified Musculoskeletal Deformities (7548)	NUMBER	69	32	4	1	5
	RATE	0.16	0.08	0.11	0.03	0.13
	Lower CI	0.12	0.06	0.03	0.00	0.04
	Upper CI	0.20	0.11	0.26	0.14	0.31
OTHER CONGENITAL ANOMALIES OF LIMBS (755)	**NUMBER**	**2007**	**1823**	**189**	**206**	**203**
	RATE	**4.66**	**4.60**	**4.97**	**5.59**	**5.42**
	Lower CI	**4.46**	**4.39**	**4.29**	**4.86**	**4.70**
	Upper CI	**4.87**	**4.81**	**5.73**	**6.41**	**6.21**
Polydactyly (7550)	NUMBER	510	557	45	64	50
	RATE	1.18	1.40	1.18	1.74	1.33
	Lower CI	1.08	1.29	0.86	1.34	0.99
	Upper CI	1.29	1.53	1.58	2.22	1.76
Syndactyly (7551)	NUMBER	396	399	40	41	55
	RATE	0.92	1.01	1.05	1.11	1.47
	Lower CI	0.83	0.91	0.75	0.80	1.11
	Upper CI	1.01	1.11	1.43	1.51	1.91
Reduction Deformities of Upper Limb (7552)	NUMBER	217	254	33	18	23
	RATE	0.50	0.64	0.87	0.49	0.61
	Lower CI	0.44	0.56	0.60	0.29	0.39
	Upper CI	0.58	0.72	1.22	0.77	0.92

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Reduction Deformities of Lower Limb (7553)	NUMBER	108	120	16	15	14
	RATE	0.25	0.30	0.42	0.41	0.37
	Lower CI	0.21	0.25	0.24	0.23	0.20
	Upper CI	0.30	0.36	0.68	0.67	0.63
Other Anomalies of Upper Limb Including Shoulder Girdle (7555)	NUMBER	160	177	27	27	20
	RATE	0.37	0.45	0.71	0.73	0.53
	Lower CI	0.32	0.38	0.47	0.48	0.33
	Upper CI	0.43	0.52	1.03	1.07	0.82
Other Anomalies of Lower Limb Including Pelvic Girdle (7556)	NUMBER	569	274	24	36	37
	RATE	1.32	0.69	0.63	0.98	0.99
	Lower CI	1.22	0.61	0.40	0.69	0.70
	Upper CI	1.43	0.78	0.94	1.35	1.36
Other Specified Anomalies of Unspecified Limb (7558)	NUMBER	45	42	4	5	4
	RATE	0.10	0.11	0.11	0.14	0.11
	Lower CI	0.08	0.08	0.03	0.04	0.03
	Upper CI	0.14	0.14	0.26	0.31	0.27
OTHER CONGENITAL MUSCULO-SKELETAL ANOMALIES (756)	**NUMBER**	**984**	**1079**	**106**	**126**	**107**
	RATE	**2.29**	**2.72**	**2.79**	**3.42**	**2.85**
	Lower CI	**2.14**	**2.56**	**2.28**	**2.85**	**2.34**
	Upper CI	**2.43**	**2.89**	**3.37**	**4.07**	**3.45**
Anomalies of Skull and Face Bones (7560)	NUMBER	298	357	38	40	36
	RATE	0.69	0.90	1.00	1.09	0.96
	Lower CI	0.62	0.81	0.71	0.78	0.67
	Upper CI	0.78	1.00	1.37	1.48	1.33
Anomalies of Spine (7561)	NUMBER	108	141	18	15	18
	RATE	0.25	0.36	0.47	0.41	0.48
	Lower CI	0.21	0.30	0.28	0.23	0.29
	Upper CI	0.30	0.42	0.75	0.67	0.76
Other Anomalies of Ribs and Sternum (7563)	NUMBER	54	89	11	12	11
	RATE	0.13	0.22	0.29	0.33	0.29
	Lower CI	0.09	0.18	0.14	0.17	0.15
	Upper CI	0.16	0.28	0.52	0.57	0.52

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Chondrodystrophy (7564)	NUMBER	54	50	4	10	3
	RATE	0.13	0.13	0.11	0.27	0.08
	Lower CI	0.09	0.09	0.03	0.13	0.02
	Upper CI	0.16	0.17	0.26	0.50	0.23
Osteodystrophies (7565)	NUMBER	34	28	3	2	2
	RATE	0.08	0.07	0.08	0.05	0.05
	Lower CI	0.05	0.05	0.02	0.01	0.01
	Upper CI	0.11	0.10	0.22	0.19	0.18
Anomalies of Diaphragm (7566)	NUMBER	170	137	15	23	18
	RATE	0.39	0.35	0.39	0.62	0.48
	Lower CI	0.34	0.29	0.22	0.40	0.29
	Upper CI	0.46	0.41	0.65	0.94	0.76
Anomalies of Abdominal Wall (7567)	NUMBER	175	179	14	22	18
	RATE	0.41	0.45	0.37	0.60	0.48
	Lower CI	0.35	0.39	0.20	0.37	0.29
	Upper CI	0.47	0.52	0.62	0.90	0.76
Other Specified Anomalies of Muscle, Tendon, Fascia and Connective Tissue (7568)	NUMBER	86	94	0	2	1
	RATE	0.20	0.24		0.05	0.03
	Lower CI	0.16	0.19		0.01	0.00
	Upper CI	0.25	0.29		0.19	0.13
CONGENITAL ANOMALIES OF THE INTEGUMENT (757)	**NUMBER**	**406**	**378**	**28**	**41**	**39**
	RATE	**0.94**	**0.95**	**0.74**	**1.11**	**1.04**
	Lower CI	**0.85**	**0.86**	**0.49**	**0.80**	**0.74**
	Upper CI	**1.04**	**1.05**	**1.06**	**1.51**	**1.42**
Hereditary Edema of Legs (7570)	NUMBER	3	3	0	3	0
	RATE	0.01	0.01		0.08	
	Lower CI	0.00	0.00		0.02	
	Upper CI	0.02	0.02		0.23	
Ichthyosis Congenita (7571)	NUMBER	20	10	1	1	2
	RATE	0.05	0.03	0.03	0.03	0.05
	Lower CI	0.03	0.01	0.00	0.00	0.01
	Upper CI	0.07	0.05	0.13	0.14	0.18

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Dermatoglyphic Anomalies (7572)	NUMBER	89	116	10	9	11
	RATE	0.21	0.29	0.26	0.24	0.29
	Lower CI	0.17	0.24	0.13	0.11	0.15
	Upper CI	0.25	0.35	0.48	0.46	0.52
Other Specified Anomalies of Skin (7573)	NUMBER	180	158	8	21	18
	RATE	0.42	0.40	0.21	0.57	0.48
	Lower CI	0.36	0.34	0.09	0.35	0.29
	Upper CI	0.48	0.47	0.41	0.87	0.76
Specified Anomalies of Hair (7574)	NUMBER	3	5	0	0	1
	RATE	0.01	0.01			0.03
	Lower CI	0.00	0.00			0.00
	Upper CI	0.02	0.03			0.13
Specified Anomalies of Nails (7575)	NUMBER	26	24	6	4	3
	RATE	0.06	0.06	0.16	0.11	0.08
	Lower CI	0.04	0.04	0.06	0.03	0.02
	Upper CI	0.09	0.09	0.34	0.27	0.23
Specified Anomalies of Breast (7576)	NUMBER	70	53	2	1	3
	RATE	0.16	0.13	0.05	0.03	0.08
	Lower CI	0.13	0.10	0.01	0.00	0.02
	Upper CI	0.21	0.17	0.18	0.14	0.23
Other Specified Anomalies of the Integument (7578)	NUMBER	12	9	1	1	1
	RATE	0.03	0.02	0.03	0.03	0.03
	Lower CI	0.01	0.01	0.00	0.00	0.00
	Upper CI	0.05	0.04	0.13	0.14	0.13
CHROMOSOMAL ANOMALIES (758)	**NUMBER**	**623**	**800**	**100**	**102**	**103**
	RATE	**1.45**	**2.02**	**2.63**	**2.77**	**2.75**
	Lower CI	**1.34**	**1.88**	**2.14**	**2.26**	**2.24**
	Upper CI	**1.57**	**2.16**	**3.20**	**3.36**	**3.33**
Down Syndrome (7580)	NUMBER	402	454	45	54	57
	RATE	0.93	1.14	1.18	1.47	1.52
	Lower CI	0.84	1.04	0.86	1.10	1.15
	Upper CI	1.03	1.26	1.58	1.91	1.97

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Patau Syndrome (7581)	NUMBER	32	46	3	3	
	RATE	0.07	0.12	0.06	0.08	0.08
	Lower CI	0.05	0.09	0.01	0.02	0.02
	Upper CI	0.10	0.15	0.34	0.23	0.23
Edwards Syndrome (7582)	NUMBER	71	104	14	13	15
	RATE	0.16	0.26	0.37	0.35	0.40
	Lower CI	0.13	0.21	0.20	0.19	0.22
	Upper CI	0.21	0.32	0.62	0.60	0.66
Autosomal Deletion Syndrome (7583)	NUMBER	24	35	8	6	5
	RATE	0.06	0.09	0.21	0.16	0.13
	Lower CI	0.04	0.06	0.09	0.06	0.04
	Upper CI	0.08	0.12	0.41	0.35	0.31
Other Conditions Due to Autosomal Anomalies (7585)	NUMBER	39	75	16	8	10
	RATE	0.09	0.19	0.42	0.22	0.27
	Lower CI	0.06	0.15	0.24	0.09	0.13
	Upper CI	0.12	0.24	0.68	0.42	0.49
Gonadal Dysgenesis (7586)	NUMBER	39	45	6	11	6
	RATE	0.09	0.11	0.16	0.30	0.16
	Lower CI	0.06	0.08	0.06	0.15	0.06
	Upper CI	0.12	0.15	0.34	0.53	0.34
Klinefelter Syndrome (7587)	NUMBER	7	18	2	4	2
	RATE	0.02	0.05	0.05	0.11	0.05
	Lower CI	0.01	0.03	0.01	0.03	0.01
	Upper CI	0.03	0.07	0.18	0.27	0.18
Other Conditions Due to Sex Chromosome Anomalies (7588)	NUMBER	7	17	3	3	5
	RATE	0.02	0.04	0.08	0.08	0.13
	Lower CI	0.01	0.03	0.02	0.02	0.04
	Upper CI	0.03	0.07	0.22	0.23	0.31
Conditions Due to Anomalies of Unspecified Chromosome (7589)	NUMBER	2	3	0	0	0
	RATE	0.00	0.01			
	Lower CI	0.00	0.00			
	Upper CI	0.02	0.02			

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
OTHER AND UNSPECIFIED CONGENITAL ANOMALIES (759)	**NUMBER**	**468**	**458**	**60**	**41**	**27**
	RATE	**1.09**	**1.16**	**1.58**	**1.11**	**0.72**
	Lower CI	**0.99**	**1.05**	**1.20**	**0.80**	**0.48**
	Upper CI	**1.19**	**1.27**	**2.03**	**1.51**	**1.05**
Anomalies of Spleen (7590)	NUMBER	41	56	11	7	4
	RATE	0.10	0.14	0.29	0.19	0.11
	Lower CI	0.07	0.11	0.14	0.08	0.03
	Upper CI	0.13	0.18	0.52	0.39	0.27
Anomalies of Adrenal Gland (7591)	NUMBER	11	8	0	1	1
	RATE	0.03	0.02		0.03	0.03
	Lower CI	0.01	0.01		0.00	0.00
	Upper CI	0.05	0.04		0.14	0.13
Anomalies of Other Endocrine Glands (7592)	NUMBER	24	27	4	1	2
	RATE	0.06	0.07	0.11	0.03	0.05
	Lower CI	0.04	0.04	0.03	0.00	0.01
	Upper CI	0.08	0.10	0.26	0.14	0.18
Situs Inversus (7593)	NUMBER	28	47	5	5	4
	RATE	0.07	0.12	0.13	0.14	0.11
	Lower CI	0.04	0.09	0.04	0.04	0.03
	Upper CI	0.09	0.16	0.30	0.31	0.27
Conjoined Twins (7594)	NUMBER	8	4	0	0	0
	RATE	0.02	0.01			
	Lower CI	0.01	0.00			
	Upper CI	0.04	0.03			
Tuberous Sclerosis (7595)	NUMBER	12	9	1	1	2
	RATE	0.03	0.02	0.03	0.03	0.05
	Lower CI	0.01	0.01	0.00	0.00	0.01
	Upper CI	0.05	0.04	0.13	0.14	0.18
Other Hamartoses Not Elsewhere Classified (7596)	NUMBER	10	4	1	0	1
	RATE	0.02	0.01	0.03		0.03
	Lower CI	0.01	0.00	0.00		0.00
	Upper CI	0.04	0.03	0.13		0.13

Diagnostic Category and ICD-9/BPA Code		80-89 Subtotal	90-99 Subtotal	1999	2000	2001
Multiple Congenital Anomalies So Described (7597)	NUMBER	24	1	0	0	0
	RATE	0.06	0.00			
	Lower CI	0.04	0.00			
	Upper CI	0.08	0.01			
Other Specified Anomalies (7598)	NUMBER	293	300	38	26	13
	RATE	0.68	0.76	1.00	0.71	0.35
	Lower CI	0.60	0.67	0.71	0.46	0.18
	Upper CI	0.76	0.85	1.37	1.03	0.59
Congenital Anomaly, Unspecified (7599)	NUMBER	18	2	0	0	0
	RATE	0.04	0.01			
	Lower CI	0.02	0.00			
	Upper CI	0.07	0.02			

Number = Defects occurring in Live Births and Stillbirths ≥ 20 weeks or ≥ 500 g CI = Approximate 95% Confidence Intervals
* = rates based on total **female** births † = rates based on total **male** births

Appendix A2. Selected Anomalies with Rates of Live Births and Stillbirths Compared with Total Rates Including Terminations of Pregnancy (ToP)

Table A2.1 Selected anomalies and rates/1000 including terminations, 1999

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	6	1	0.16	0.18
Spina Bifida	13	2	0.34	0.39
Encephalocele	2	3	0.05	0.13
Hydrocephaly	19	0	0.50	0.50
Digestive System (*751..)	104	16	2.73	3.15
Urinary System (*753..)	162	14	4.26	4.62
Limb Anomalies (*755..)	189	21	4.97	5.51
Musculo-skeletal Anomalies (*756..)	106	16	2.79	3.20
Chromosome Anomalies (all)	100	33	2.63	3.49
Down Syndrome	45	20	1.18	1.71
Syndromes	38	5	1.00	1.13

*numbers indicate ICD-9/BPA code

Table A2.2 Selected anomalies and rates/1000 including terminations, 2000

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	8	4	0.22	0.33
Spina Bifida	7	2	0.19	0.24
Encephalocele	4	0	0.11	0.11
Hydrocephaly	27	3	0.73	0.81
Digestive System (*751..)	76	12	2.06	2.39
Urinary System (*753..)	144	10	3.91	4.17
Limb Anomalies (*755..)	206	14	5.59	5.96
Musculo-skeletal Anomalies (*756..)	126	16	3.42	3.85
Chromosome Anomalies (all)	102	31	2.77	3.61
Down Syndrome	54	11	1.47	1.76
Syndromes	26	2	0.71	0.76

*numbers indicate ICD-9/BPA code

Table A2.3 Selected anomalies and rates/1000 including terminations, 2001

Congenital Anomalies (CAs)	Number of Anomalies		Rates/1000 Total Births	
	Live (L) and Still (S)	ToP (T)	L + S	L + S + T
Anencephaly	3	4	0.08	0.19
Spina Bifida	11	1	0.29	0.32
Encephalocele	6	0	0.16	0.16
Hydrocephaly	21	3	0.56	0.64
Digestive System (*751..)	74	11	1.97	2.26
Urinary System (*753..)	176	21	4.70	5.25
Limb Anomalies (*755..)	203	18	5.42	5.89
Musculo-skeletal Anomalies (*756..)	107	16	2.85	3.28
Chromosome Anomalies (all)	103	23	2.75	3.36
Down Syndrome	57	14	1.52	1.86
Syndromes	13	5	0.35	0.48

*numbers indicate ICD-9/BPA code
ToP = Termination of pregnancy

Appendix A3. Numbers of Cases, Anomalies and Anomalies per Case

Table A3.1 Numbers of cases, anomalies and anomalies per case 1980-2001

Year	Total births (Live and Still)	Cases (Live and Still)	Case Rate per 1000 Total births	Number of Anomalies (Live and Still)	Anomaly Rate per 1000 Total births	Average Anomalies per case
1980	39,655	1,423	35.88	1,874	47.26	1.32
1981	42,463	1,517	35.73	2,041	48.65	1.35
1982	44,987	1,625	36.12	2,224	49.44	1.34
1983	45,381	1,543	34.00	2,202	48.52	1.43
1984	43,864	1,623	37.00	2,229	50.82	1.37
1985	43,565	1,693	38.86	2,423	55.62	1.43
1986	43,552	1,780	40.87	2,445	56.14	1.37
1987	41,957	1,696	40.42	2,439	58.13	1.44
1988	41,970	1,874	44.65	2,731	65.07	1.46
1989	43,223	1,933	44.72	2,863	66.24	1.48
1990	42,895	1,985	46.28	2,947	68.70	1.48
1991	42,675	1,780	41.71	2,556	59.89	1.44
1992	41,944	1,775	42.32	2,599	61.96	1.46
1993	40,163	1,469	35.29	2,167	53.96	1.48
1994	39,720	1,409	35.47	2,127	53.55	1.51
1995	38,784	1,179	30.40	1,809	46.64	1.53
1996	37,710	1,141	30.26	1,746	46.30	1.53
1997	36,798	1,079	29.32	1,805	49.05	1.67
1998	37,808	1,139	30.13	1,854	49.04	1.63
1999	38,026	1,166	30.66	2,166	56.96	1.86
2000	36,839	1,249	33.90	2,098	56.95	1.68
2001	37,484	1,327	35.40	2,208	58.91	1.66
Total	**901,463**	**33,405**	**37.06**	**49,553**	**54.97**	**1.48**

Appendix A4. Termination of Pregnancy – Anomalies per Case

Table A4.1 Termination of pregnancy (ToP) not registered with Alberta Vital Statistics (gestation <20 weeks or birth weight <500g)

Year	# ToP cases	# ToP anomalies	Average Number of Anomalies/Case
1997	65	126	1.94
1998	70	232	3.31
1999	54	185	3.43
2000	53	148	2.79
2001	52	175	3.37
Total	294	866	2.95